# Familial complex tics and autistic behaviour with favourable outcome in young children

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SUMMARY

The author presents in this article, through her experience with children with autism from the 80s up to the present data, the presence of complex tics in children and their families.

KEYWORDS

Tics, autism.

## Introduction

Young children with autistic behaviour and subsequent relevant improvement with disappearance of autistic series symptoms and appearance of normal mental abilities have been reported by various authors (Lovacs, 1987– McEachin *et al.*, 1993; Perry *et al.*, 1995), including parents (Rimland. 1994), and represent an object of controversy in the profession. Some of these children subsequently reveal the development of other pathologies such as mood disorders, social phobia, etc. (Zappella, 1996) but remain free from the features of mental retardation and autism.

In the course of my experience with children with autism, I have been able to observe some of these children from the beginning of the 80s and to follow them up to the present date. More recently, I was able to identify the presence of complex tics (motor and, sometimes, vocal) in a number of these subjects, as well as in other members of their family. This neurobiologic feature together with other elements observed during follow up suggest the possibility that these children represent a particular subgroup within the autistic spectrum. In addition, other questions remain unanswered, and among them three appear relevant: 1) what is the long term outcome, 2) how many are they, compaired to a usual population of children with autistic behaviour, 3) do they represent a special subgroup within the autistic spectrum disorders, and should they be treated according to special criteria?

The present article attempts to contribute to an appropriate answer to these questions both reporting some cases followed over many years, and describing an outpatient population observed in the south of Italy.

## Materials and method

Among the patients who over the years have attended the Department of Child Neuropsychiatry in Siena, two subjects are described who had a diagnosis of autism in early 80's and subsequently mantained contact with the Department. These subjects are described as Cases 1 and 2 and were treated with a variant of the holding therapies coupled with family therapy (Zappella, 1987).

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In addition, an outpatient population, which the present author has been attending in South of Italy (Calabria) over the last three years, has also been screened, taking into account the total number of children with autism observed with particular reference to those who underwent a pronounced steady improvement and with disappearance of the autistic features and appearance of normal mental abilities. All the children were evaluated by the author clinically with the aid of DSM-IV criteria.

A family history of psychiatric disorders was also obtained with particular attention to mood and anxiety disorders as well as to complex tics over three generations.

Their abilities were identified with the checklist of the Portage method, both during the initial and the final evaluation. Cases of classic Rett syndrome were excluded but cases of the preserved speech variant (Zappella, 1997) were included in the series.

Cases with Asperger syndrome were also included. Children belonging to this series were treated with AERC (Zappella, 1996).

AERC is based on supporting reciprocal corporeal and emotional interactions between the child and one of his parents along lines of primary intersubjectivity. It includes motor activation of the child, inducing fun and amusement, followed by cooperation in the lines of secondary intersubjectivity. It favours communication at a preverbal, verbal and gestural level. It has an ethological foundation which suggests that some systems of movements (i.e. affiliation, attachment, friendliness, exploration of the other, etc.) belonging to the same larger group (approach behaviour) may reinforce each other. These theoretical assumptions explain the rapid improvements that can be obtained in some children with difficulties in their maturation. It is often implemented by the use of the checklist of the Portage method which represents a useful tool for helping parents to guide their children in resuming appropriate abilities.

## Results

The outpatient population observed in Calabria was represented by 93 children with autism, 63 of whom were males and 20 females. Age range at the first visit was between 3 and 16 years (mean age 7 years 7 months). Among these children there were three couples of twins (two male and one female) and two sisters of different age. Two children had very limited sight and in one case rubella during pregnancy was the relevant etiologic factor. Two of the males had Asperger syndrome. Among the girls, three were diagnosed as examples of the preserved speech variant of the Rett complex.

This series can also be split considering the subgroup of those between 3 and 5 years of age: a total number of 33 children, 29 males and 4 females. Females are apparently underrepresented in the younger age group. It must be noticed that, even in those children with a more favourable outcome, parents tended to hide the presence of complex tics in themselves or other relatives and this was revealed at times, only after repeated enquiries, in subsequent visits. It may well be therefore that the presence of this disorder is underrepresented in those children with a less favourable outcome.

Only 4 male children, initially observed between 3 and 5 years, had a favourable outcome, completely shedding their autistic features and resuming normal or close to normal abilities: 4.3% in the general series, 12.1% in the 3–5 year series. In all of them familial complex tics were present (see Table 1). One of these cases, case A, is described in detail. Two other cases (5 and 6) shared a similar familial background with the presence of onicophagia and complex motor tics in relatives belonging to the second generation: however, the outcome of the child was autism and normal abilities were not resumed.

## Case reports

### Case 1

The youngest of three brothers. His father had complex motor tics of the face and his mother suffered from depressive episodes prior to his birth. He was the result of a normal pregnancy and delivery. His development is reported as normal up to 10 months when he was beginning to utter a couple of words. At that age he had a febrile convulsion which unduly alarmed his mother. In the subsequent months he had some occasional blue spasm and at the same time his behaviour began to deteriorate: he began actively avoiding interactions with other people and he would occasionally interact with their requests by beating his head against the wall or on the floor; he would not react if called by name and used objects repetitively turning them over in his hands. At 1 year 10 months he had a normal EEG. Nevertheless a treatment was started with phenobarbital and valproate.

His mental conditions did not change in the following year when a diagnosis of autism was made by a specialist in child neuropsychiatry. At 3 years and a half he was examined for the first time by me in the outpatient's section of this department: he was unable to speak, did not play with objects, was unable to put one cube on to the other and was entirely uninterested in dolls or puppets: he showed a number of stereotypic

**TABLE 1**

| | A | B | C | D | E | F |
|---|---|---|---|---|---|---|
| I | 3y | 5y | 2y10m | 3y6m | 4y | 6y6m |
| II | 5y | 6y | 3y10m | 4y4m | 4y8m | 5y10m |
| III | 2y | 1y | 1y | 10m | 8m | 4m |
| IV | 18m | 4y | 2yl0m | 2y | 2–3y | 2–3y |
| V | 5y6m | 5y | 3y10m | 4y4m | 4y | 2–3y |
| VI | (1)4(2)3 | (1)1(2)1(3)3 | 1(2)1(3)1 | (1)2(3)1(3)1 | (1)2(2)2 | (1)2(2)3(3)2 |
| VII | | 0 | 0 | 0 | 0 (1)3(2)3(3)4 | (1)1(2)2(3)2 |
| VIII | CMT&VT | CMT | CMT | CMT | CMT | CMT |
| IX | Ma, Pu, Pc | Pu | 2Mu, M | F, Pu, M, Ma | F, 3Pu, M, Mu | Ma |
| X | 3 | 3 | 2 | 2 | 4 | 3 |
| XI | 6 | 4 | 3 | 5 | 2 | 2 |
| XII | DD | in PU | | | | |

*Legend for Table 1*
I: age at the first visit
II: age at the last visit
III: follow up in years (y) and months (m)
IV: mental abilities at the first visit
V: mental abilities at the last visit
VI: DSM-IV values at the first visit
VII: DSM-IV values at the last visit
VIII: CMT (Complex motor tics); VT (vocal tics)
IX: M (mother), F (father), Ma (maternal aunt), Pa (paternal aunt), Mu (maternal uncle), Pu (paternal uncle)
X: total number of brothers and sisters of the father
XI: total number of brothers and mothers of the mother
XII: other pathologies within the two generations, DD (depressive disorder)

activities with his hands, avoided eye contact and in general was a rather withdrawm child who passed most of his time alone, interrupting his loneliness occasionally, for example, throwing himself over a table and destroying all the objects lying on it. From a neurological point of view he appeared otherwise to be normal. Speaking with his parents it was evident that his mother was depressed and a treatment was therefore started having his father hold him in his laps in a face to face interaction, speaking to him as if he were a baby. The child reacted with rage at first but subsequently accepted this type of interaction more willingly. At the end of the session which lasted half an hour there was a free play interaction between the child and his father. The family was dismissed with the indication to practice these sessions every day. It was asked to come to our outpatient's department every month. The following month the child had started to say a few words and was showing new signs of affection towards his mother. At the third session his abilities were further increased: he was able to built a tower of nine cubes, was able to point to various objects, to say a few more words and showed an improved understanding.

Phenobarbital and subsequently valproate were slowly withdrawn. At the sixth visit it was evident that the holding, which in the last four months had been conducted only occasionally, was an object of controversy at home and I advised the family to interrupt it, giving in its place some advise aimed at improving family structure and communication. At that time the child had already been attending a nursery for about a month. In the following session, he was much more ready to cooperate. The parents were then instructed to teach him new activities: regular improvement ensued in the following months but his speech was still frequently echolalic by 4 years and a half. At this age he started speech therapy and after an interval of six months he was seen again at 5 years 8 months. It was clear that there were no more elements of autistic behaviour and his mental abilities were within the normal limits. He was able to engage in make believe plays in a way adequate to his age, and interacted well with his pairs.

In subsequent years T. had a normal scholastic curriculum up to the end of the high school. Afterwards he did military service for a year and is now following a vocational course in order to work in the field of tourism.

I met him a month ago and found no elements of autism with DSM-IV evaluation: he is a normal, pleasant boy with several friends and the usual activities of 19 year old boys. He still mantains complex motor tics with extension of the arms and squeezing of the fingers, tics which become more intense when he is excited.

### *Case 2*

There is a history of major depression episodes in his mother prior to the birth of this child and in both his maternal grand parents. His mother has motor complex tic of her hands, a maternal aunt has onicophagia and his father has motor complex tics (extending both arms and squeezing his fingers). He is the second of two children. Pregnancy and delivery were normal. His initial development was normal: he was able to walk alone and to say his first words at 11 months but during his second year of life there was a regression, he lost the few words he had and was subsequently diagnosed as affected by autism, I first saw him at 3 years 11 months of age. He avoided interactions, and engaged in repetitive activities with his hands, 'squeezing' his fingers, much like his father, and was frequently seen turning a ball or other objects over themselves, was unable to draw, was able to put a few cubes one into another to make a tower and could say only one word. A holding session was started with his mother, and the child soon began to interact better and say a few more words. His subsequent treatment continued with a trend similar to case 1: a combination of holding interactions, conducted by his mother, family therapy and pedagogic indications: there were improvements and short regressions in the first months but subsequently improvement was more regular with some echolalia still present at 5 years: however, by the age of 5 years 8 months he showed no more signs of autistic behaviour, and normal mental abilities.

At this age, and for 5 years a complex facial motor tic was present (as it had been in his father when younger) which lasted about 6 years. He attended school regularly up to 15 years old. In his adolescence he was not popular with his peers and was frequently teased, and had some oppositional disagreements with his father. More recently he has become more integrated with his peers and has been dating some girls. He was seen for the last time at 18 when a WISC showed an I.Q. of 98. There were no elements suggesting autism in his evaluation. His social difficulties were minor and could have a number of environmental explanations, including the fact of living in a very small and isolated village. Otherwise he is a nice young man, adequate in his ability to recall past events, to begin and sustain a conversation and to identify people's mental states, not particularly different from other boys living in similar social surroundings.

Case 3. He is an only child. His family history includes a paternal cousin with episodes of major depression, a paternal uncle and his son with complex motor tics and a maternal aunt with onicophagia. Pregnancy and delivery were uneventful. His initial development was normal and at 1 year 1 month he was able to walk alone. At 2 years he started to say his first words: at that age, he appeared particularly hyperactive and when playing with puppets merely lined them up but not in a metaphorical, fiction way. In the subsequent year he increased his vocabulary to a modest degree but was unable to interact or play with his peers and never had developed a symbolic play: around 2 years and a half his mother noticed a certain degree of regression and loneliness. A diagnosis of autism was therefore suggested. He was examined by myself at 3 years and appeared to be an avoiding child with poor expressive abilities, who showed no elements of joint attention, nor reciprocal socio-emotional interaction, did no gesture and displayed an idiosyncratic speech, limited to short phrases and single words. DSM-IV for autism showed (1) a+, b+, c+, d+(2) a+, b+, c+, d+(3)c+. Parents were advised to: 1. Have frequent reciprocal corporeal interactions with him, inducing fun and amusement, face to face, activate him on a motor point of view (AERC, see Zappella, 1996; 1998) and to guide him to gain new abilities with the help of the Portage checklist and method. Speech therapy was also advised as well as send him in a nursery. This child was followed carefully along these lines and was seen subsequently as an outpatient on three occasions. At 4 years he developed simple ocular tics and facial motor complex tics which remained in the following 12 months. At 5 years the child appeared entirely normal and of apparently brilliant intelligence: his reading was good and he was able to write and count up to 30. Except for (3), positive because of the persistance of sonic complex motor tics, no other item of DSM-IV was positive. He was able to speak adequately, to tell stories, to use make believe in play with his peers, appropriate to his age. The child, however, appeared hyperactive and impulsive and fullfilled the criteria of ADHD.

These children had no dysmorphic features, their neurological evaluation was normal as well as their head circumference.

## *Discussion*

The present data support the notion of the existence within the autistic spectrum, of a subgroup with peculiar neurobiologic dysmaturational features and familial complex tics (motor and vocal) compatible with a favourable outcome already described in ten cases (Zappella, 1994; 1996). The neurobiological profile of these children may include different variants: case 3

turned out in the end to be a probable ADHD and his behaviour, described around two and a half years, was similar to what at times occurs in some young children with ADHD: he was less responsive and isolated than before. In addition, he lagged behind in symbolic play and social abilities. This picture is not the same as in case 1 and 2, where profound regression was in clear contrast with previously normal development. Case 2 had undergone clear regression by 2 years old when he was diagnosed as autistic in an University Centre: when he was subsequently seen by myself he initially manifested dysmaturational elements such as difficulties in oral and manual coordination, was easily afraid of the wind, the dark, showers, unknown people and situations) and used to wipe away saliva if kissed. Case 1 had also undergone a clear regression at one and a half year and was diagnosed as autistic at 2 years in a specialized hospital: in this case clear dysmaturational features as in Case 2 were not evident. But Case 2 developed normally in the first year of life, however by his second year he appeared to be a lonely child and was not engaging in functional play. He was seen at 3 years 10 months when he dysplayed a full autistic picture (see Table 1), with a number of motor and vocal tics already, in addition he was hyperactive. 10 months later he had completely recovered: no more signs of autistic behaviour could be detected, he interacted well with his peers, was able to use make believe in play adequate to his age and was already able to read.

The clinical picture of these children included normal development in the first year of life, normal motor milestones (all of them walked before 14 months), normal head circumference but at a certain period of their life below two and a half years autistic behaviour was present which was directly observed by myself or reported by specialized centres. In most of them a full picture of autistic behaviour appeared as part of a regression. A notable feature was their rapid improvement following intense, reciprocal, corporeal interaction with or without motor activation, often from their first visit: this fact, together with the observation of familial complex tics, the absence of dysmorphic elements, the presence in some cases of dysmaturational features (sensory disturbances, delayed motor coordination) led to the promise of a favourable outcome. Rapid recovery in the following visit gave further support to this hypothesis. It must be stressed, however, that these elements can only suggest a possible favourable outcome: there are in fact children with all the above quoted constellation of symptoms who remain autistic or otherwise handicapped. Familial complex tics can in fact be accompanied in some cases by persistent Autism in the young proband, as in cases 5 and 6 of the present series, who were treated with the same therapeutic approach as those with a more favourable development: in these cases follow up showed the persistance of autistic behaviour.

Cases 1 and 2 were observed 15 years ago and, although the symptoms of an autistic disorder were well documented at that time, the neurodevelopmental evaluation was less rich in details than in the recently observed series. When seen in their late teens they had no elements of Autism. They were both boys of average intelligence with social abilities falling within the normal range. When given the questions used for Asperger syndrome (Frith, 1991), their answers were normal.

The neurobiological basis of this developmental disorder may include different variants and the frequent presence of familial complex tics can go together with ADHD or with dysmaturational elements of a different kind: in similar but eventually not coincident ways these children may become more vulnerable to environmental difficulties which were documented in some cases. In case C there was a marked conflict between parents, both overdependent on their respective family of origin, and in case 1 regression appeared to go together with the emotional state of an overanxious and depressed mother. It may well be that the sensory and perceptual disorder and the delayed praxic abilities, which are clearly documented in some children, contribute in a significant way to their interactional difficulties and make these children more sensitive even to minor disorders of their surroundings.

The cases presented in this article were treated, respectively, with a variant of the holding therapy coupled with family therapy (Zappella, 1984; 1987) and with AERC. These therapeutic approaches have something in common: the holding method used with Cases 1 and 2 included a reciprocal corporeal, emotional and spoken face to face interaction, followed by parental teaching of new abilities. In this way the interactional tools of babies were intensively proposed (primary intersubjectivity, Trevarthen, 1980) followed by proposals of cooperation. This approach had, however, some notable disadvantages and it appears archaic in the light of present knowledge: the child's legs were kept under constraint for considerable periods of time, contrasting with the increased close facial interaction, often causing a rage response which disturbed the child and his parent (Zappella, 1998). In the cases observed in recent years primary and secondary intersubjectivity were also powerfully proposed together with motor activation in a dimension based essentially on evoking pleasure and fun (AERC, Zappella, 1996): improvement was apparently more rapid and obtained with on relevant stress on behalf of the child and his parents.

Prekop in 1984, using the holding technique, also found a favourable outcome in a percentage of her cases:

it is interesting to note that the Prekop series had similar percentages of normal outcomes (13 out of 104 cases) as in the series reported by myself (6 out of 50 cases, i.e. 12%) both using a similar approach at that time (Zappella, 1987). It therefore seems that different therapeutic techniques like holding and AERC which have in common a powerful intervention on reciprocal interactions along the lines of normal development (i.e. primary and secondary intersubjectivity) give positive results in a similar percentage of young children with autistic disturbance corresponding to an approximate 10% in a population of young children with autistic behaviour seen by a specialist working on these disorders. A favourable outcome for young children with autistic behaviour has also been reported by Lovaas (1987), McEachin *et al.* (1993) and by Perry *et al.* (1995) through operant conditioning and it must be noticed that this therapeutic approach includes an intensive face to face interaction, often accompanied by a warm, supportive attitude and periods of relaxation. In other words, in spite of notable differences with the above mentioned therapies, primary and secondary intersubjectivity are also intensively proposed to the child with the operant conditioning therapy.

Greenspan and Wieder (1997) have recently described the outcome of young children with autistic disturbances treated with an interactional therapy named 'floor therapy', which is also active in terms of primary and secondary intersubjectivity, and described similar disorders of maturation in some of those with a more favourable outcome, labelling them as multisystem developmental disorder in order to distinguish them from such devastating disorders as most autistic syndromes. These authors mention that 58% of their series lost autistic features with CARS but they do not state the developmental outcome of their children with other replicable scales. In short it is possible to say that a number of young children with autistic disturbances are in strong need of resuming communication along the lines of primary and secondary intersubjectivity with a more powerful and intense emotional interaction. A few, definite methods can contribute to a positive outcome. Among them the AERC approach is by far the more simple and economic as it requires a limited number of sessions (a total of three to four for every child seen in this outpatient's series).

Few wonder if other therapeutic approaches where the lines of normal developmental interaction are not intensively followed and, in contrast, a rigid temporo-spatial structured series of monotonous activities are proposed in unnatural settings such as special schools cannot claim any positive result of this kind. These and many other approaches (see for a review Howlin, 1997), when confronted with children of the kind described in this article, share three elements which are misleading for these cases: a definition of autistic behaviour in young children coincident with the diagnosis of autism as an incurable condition, the use of the therapeutic method in a separate, special institutional or scholastic context, often including only children with autism and therapeutic strategies which are not consistent with the lines of normal interactional development. It is possible that these therapies lead to a self-fulfilling profecy in which the professional expectations are confirmed by the unfavourable outcome.

RESUMO

A autora demonstra neste artigo, sua experiência com crianças autistas desde os anos 80 até os dias atuais, identificando a presença de tiques complexos tanto em crianças quanto em outros membros da família.

PALAVRAS-CHAVE

Tiques, autismo.

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